10

A CASE OF CHLOROMA.[1]

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WITH ADDITIONAL MICROSCOPICAL EXAMINATION AND PHOTOGRAPH.

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IT IS possible that the subject of chloroma is as new and rare to many of you as it was to me when the case I have to report came into my hands. It only came into my service at the Ophthalmic Hospital because the neoplasm had invaded the orbits and produced a marked exophthalmus. In the *American Journal of the Medical Sciences* for August, 1893, appears a paper entitled "Chloroma and Its Relation to Leukæmia," by Dr. George Dock, of Michigan, in which he reports a case. He has examined the literature of all languages and collected, including his own, seventeen cases of chloroma.

Edward Ludwig, aged 7 years, was admitted to the Ophthalmic Hospital May 18, 1893. His parents are living and in good health. There are five other children in the family, all of whom are healthy.

PREVIOUS HISTORY.—He has always been a bright, intelligent boy and has never suffered from any serious illness. He has had repeated attacks of parotitis (?) and the present trouble seems to have followed one of these attacks. He at first complained of pain in both orbital regions, at the same time both eyes seemed to be more than usually prominent. After a week or more, his mother noticed some impairment of his hearing, which slowly increased The exophthalmus increased more rapidly. He became very weak and somewhat emaciated, and complained of a severe pain in the left foot, on account of which he was unable to walk.

Upon his admission into the hospital both globes were very prominent, but the right more so than the left. It was with difficulty that he could close the lids of the right eye over the ball. The corneæ are clear and there is no impairment of vision. The conjunctivæ of both eyes are red and chemotic. The motility of the eyes is unimpaired. In

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[1] We republish this paper, read before the Section of Ophthalmology of the American Medical Association, May, 1896, and published in the *Journal of the American Medical Association* of November 7, 1896, at the express desire of our esteemed collaborator, as he hopes it will thus reach more readily a public to which it must be of especial interest and since a microscopic examination of parts of the tumor was offered us.—[EDITOR]

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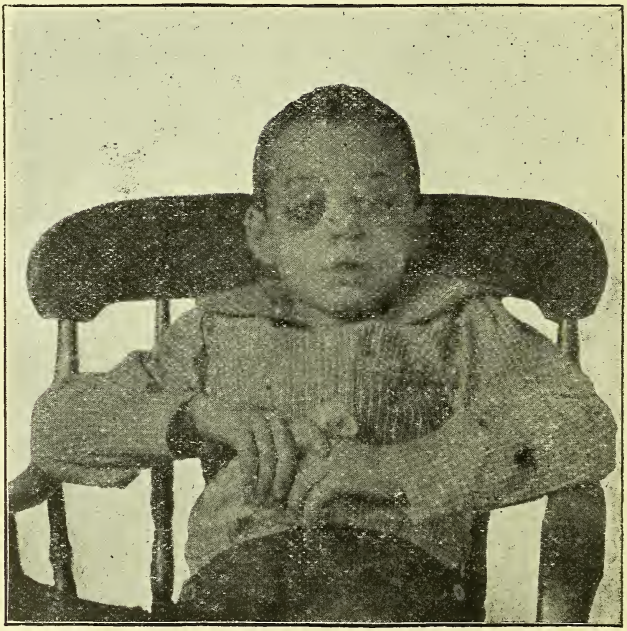
the upper and outer portion of the right orbit can be felt a dense, inelastic growth which protrudes beyond the edge of the bone. A similar growth can also be felt in the left orbit, but not so pronounced. The optic papillæ are somewhat pale, but there are no other alterations in the fundus of either eye. His hearing is so much impaired that he has to be spoken to in a loud tone. Pulse, 120 per minute; temperature, normal. There is no lesion of any of the cranial nerves. Muscular power and sensation of both upper and lower extremities normal. Pressure on the sole of left foot produces pain, but there is no redness or swelling in this locality.



May 21. The proptosis of the right eye has increased, but that of the left is apparently slightly less. There has developed a swelling in both temporal regions. They are rounded and elastic, without any sensation of fluctuation.

May 23 Protrusion of both eyes increased; corneæ hazy and covered with inspissated mucus.

May 27. Temperature, normal; pulse, 120. Patient complains of some difficulty of micturition; later on, retention of urine. Both corneæ are quite opaque. The swellings in the temporal regions more pronounced. There has developed a hard, firm swelling in the region of each parotid gland. Intelligence good; hearing somewhat improved. Pain in foot still present. No appetite. The exophthalmus of both eyes increases day by day. Both globes are now forced beyond the palpebral fissure.

June 2. Condition worse in all respects. He has had a severe hæmorrhage from the conjunctiva of the right eye.

June 5. Died this morning as result of hæmorrhage from the conjunctiva of the right eye.

Three days before death, blood pale and watery. Staining according to the Ehrlich method showed a marked increase in the number of white blood corpuscles, the increase being due principally to the number of multinuclear cells, while the small lymphocytes and those white cells characteristic of splenic and myelogenic leukæmia are not increased. Eosinophile cells are sparingly present, not above 2 per cent. The examination of the blood at this time justifies the diagnosis of leucocytosis only, and not leukæmia. Furthermore, an examination of the patient revealed no enlargement of the lymphatic glands except those of the neck.

The post-mortem examination was made about twenty-four hours after death: Body fairly well developed and nourished; post-mortem rigidity well marked, the upper and lower extremities normal. No evidence of subcutaneous hæmorrhages. Marked swelling of the lymphatic glands about the neck. Both eyeballs protrude to such an extent that they are are only partly covered by the eyelids; sloughing of both corneæ. Traces of blood about the eyes, mouth and nose. Mucous membrane of mouth swollen; teeth covered with blood. To the touch the lymphatic glands are firm and resisting, with no sign of fluctuation. A small nodular infiltration over the right parietal bone near the median line. On cutting through this swelling it was found to be situated beneath the periosteum, and although there was no evidence of its having arisen from the bone, the latter was at this point remarkably thin. No trace of inflammation or infiltration of the pia mater or the brain. The brain was found to be intensely anæmic. On removing the brain the orbital plates were seen to bulge upward owing to the pressure from within. After removing the very thin orbital plates, both cavities were found filled with a firm, solid, and somewhat elastic mass of a light greenish color, which pushed the eyeball forward. This mass was removed with some difficulty, although only at one point did it seem to be firmly adherent to the periosteum lining the orbital cavity. Here the bone was somewhat eroded. Behind, the new growth extended to the sphenoidal fissure, surrounding but not involving the optic nerve. In front and above, the newly formed tissue extended to and a little beyond the orbital arch. On superficial examination the new growth seemed to be confined to the orbital cavities; a further examination showed this not to be the case, for the cancellous tissue of the sphenoid and the petrous portion of the temporal bones were softened and infiltrated with a dirty greenish-yellow fluid, looking very much like pus. On the left side in the petrous portion of the temporal bone was an area in which the bone tissue was completely destroyed and filled up with a material such as was found in the orbits, differing from it only being of a more dirty greenish-yellow color, and not quite so firm. The lymphatic glands presented the same appearance in section as did the tumors of the orbits.

Equally as interesting proved the further examination of the new growth. On having placed the tumor in alcohol, the color disappeared entirely within twelve hours. On the other hand, the specimen kept within a tightly corked bottle, still retains the greenish color; in fact, the color has become even more marked. Some of the fresh fluid taken from the interior of the sphenoid bone was examined, but revealed only the presence of large numbers of small corpuscles, fat globules and granular detritus.

The section of the tumor after being hardened in alcohol and ether seemed to justify the diagnosis of sarcoma, the cells being almost entirely of the small round variety, none or very few spindle cells, and remarkably little intercellular substance; no large blood vessels, but numerous lymph spaces. Furthermore, the small round-celled infiltration extended into the upper eyelid down to the yellow elastic cartilage.

The seat of the tumor, the peculiar color, the history of the growth, the remarkable involvement of the lymphatic glands, the results of the microscopic examination, make this case one of a few that have been put on record under the head of chloroma, cancer vert, the green cancer of the French.

Whether the form of tumor should be called a sarcoma, whether it is of the so-called connective tissue variety, or on the other hand, as Dock would seem to believe, a peculiar condition arising in the course of true leukæmia, the tumors being regarded in this case as lymphomata—these are questions which the future must decide. Dock and a number of others have found by an examination of the blood that they were contending with true leukæmia. My examination of the the blood, as already described, showed a remarkable increase of white blood corpuscles, but they were indicative only of a marked leucocytosis, and not a true leukæmia. This examination was made three days before death. However, there is still a great deal of obscurity about leukæmia as to its true nature, and when we consider that some writers have suggested the name of sarcoma of the blood for this disease, we can not wonder that in the case of chloroma the pathologic conditions found were suggestive of leukæmia.

I am greatly indebted to Dr. Greiwe for the above report of the post-mortem and of his microscopic examination of the growths. His views, while they may differ from other investigators', are founded on examinations made of the neoplasm removed from different localities. It is very unfortunate that the post-mortem did not include the thoracic and abdominal cavities. A more complete examination might have thrown some light on the tissues invaded by this strange disease.

In all the prominent diagnostic points my case seems to correspond with the one reported by Dr. Dock, as well as most of those he has so carefully collected.

COLOR.—In color it was a yellowish-green, or perhaps a pea-green. It did not vary much in the different localities, but was more pronounced in the larger masses.

CONSISTENCY.—This differed in different localities. It was homogeneous and without fibrous septa. I have described the cut masses as presenting a smooth, shining jelly-like appearance.

INVOLVEMENT OF PERIOSTEUM AND BONE.—Most of the cases show involvement of the periosteum, and in this case it was marked, but more than this an actual softening of the bone. Dr. Greiwe in his report says that "the cancellous tissue of the sphenoid and the petrous portion of the temporal bones were softened and infiltrated with a dirty greenish-yellow fluid, looking very much like pus." On the left side in the petrous portion of the temporal bone was an area in which the bone tissue was completely destroyed and filled up with a material such as was found in the orbits, differing from it only by being of a more dirty greenish-yellow color, and not quite so firm.

DURATION.—From the report of the mother it seems that she noticed the exophthalmus only four weeks before she brought him to the hospital. He died in two weeks after he was first seen. Allowing two weeks to elapse in which the changes were not detected by the parents, the disease ran its course in the short period of two months. After he was first seen the growth was exceedingly rapid and could be noticed day by day.

HÆMORRHAGE.—Epistaxis is mentioned as occurring in some of the cases of chloroma reported. In Ludwig's case there was severe hæmorrhage from the conjunctiva of the right eye. This occurred in my patient the day before he died, and then on the following day recurred with such violence as to destroy his life.

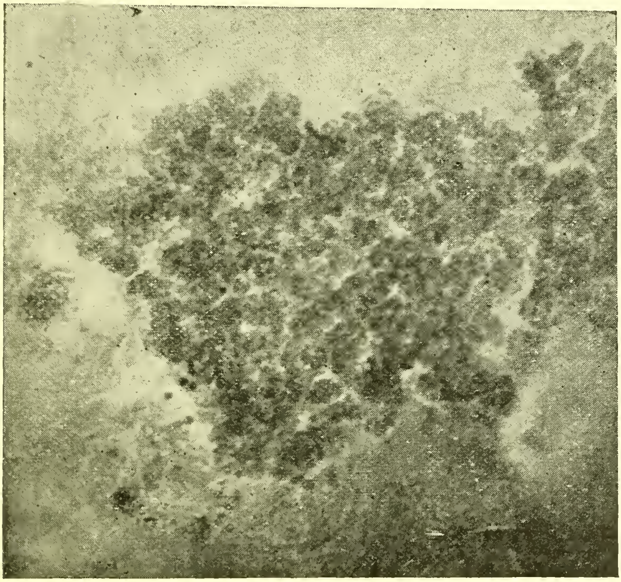
RETINA.—The eyes were carefully examined and no swelling of the optic discs and no retinitis was found. The discs were pale and anæmic, but not atrophied. His vision was good. In a short time the cornea became so involved that further inspection of the fundus was impossible.

EXOPHTHALMUS.—This was pronounced and was the first symptom that attracted the attention of his parents. The exophthalmus increased slowly in both eyes, but more in the right, and in a week from the time he was first seen the proptosis was so great that he could not close the eyelids, and in consequence the cornen ulcerated.

EXCITING CAUSES.—There is no known exciting cause in this case, but his illness followed an attack of parotitis. His mother said he had had several attacks of so-called parotitis. It is not probable that these attacks were true mumps, but precursors of the development of the disease.

DEAFNESS.—Deafness was a marked feature. It varied slightly in severity from day to day, and it was always necessary to speak to him in a loud tone. He had no catarrh and there was no otorrhœa The drum membranes were not examined.

Dr. Dock says: "From what we do know of the natural history of leukæmia and of chloroma we can see the analogies of the diseases. We can say that chloroma is a lymphomatous process similar in its clinical features to leukæmia and pseudoleukæmia. Our ignorance of its precise relations depends partly on the fact that our knowledge of chloroma is very incomplete, partly on the indefinite nature of leukæmia and pseudoleukæmia."



RESULTS OF THE MICROSCOPICAL EXAMINATION.

The parts of the tumor kindly sent me by Dr. Ayres were kept in a formol solution. They consisted of two pieces of the growth one larger and more consistent than the other. Both were of dark grass-green color. The preserving fluid also was dirty, greenish, and looked fatty.

After proper embedding I made sections of the larger and more solid-appearing piece.

In unstained sections the tissue is found to consist chiefly of small and larger round elements containing fat globules, although in one part of the tumor fibrous tissue predominates. The round elements seem to lie rather loosely together and are held by an intercellular substance which is hardly visible. Between these round cells which all have a greenish tint there lie aggregations of varying sizes composed of larger and darker bodies which are totally filled with or altogether consist of small fat or oil globules. Their shape is not exactly similar to any cell. These bodies are much more numerous in some parts of the tissue than in others. (See Figure).

The tissue takes up all the different kinds of staining materials, but poorly, nuclei can not be demonstrated and it does not photograph well therefore. Whether this is, as I should take it, a sign of partial disintegration of the tissue elements, or whether this is characteristic of the chloroma and due to the fat globules, I do not know. Yet, the latter is not probable, since the large bodies, more particularly filled with fat globules, take up more stain than the rest of the tissue.

When examining the preserving fluid in which the pieces of the tumor reached me, I found besides innumerable such bodies with fat globules the whole green fluid pervaded by octahedric crystals of perfect purity, but varying in size. They are in no way different from crystals of oxalate of calcium as we find them in the urine. When I allowed the fluid to evaporate, larger droplets of a dark green oily substance and these crystals filled the field.

Ziegler says: "Chloroma is a round cell sarcoma very rich in cells, characterized by a light green or dirty brown-green color and which, according to our present knowledge, springs from the cranial periosteum. According to Huber and Chiari the coloring substance resides in small globules contained in the cells, which give the microscopical reactions of fat. I can confirm these statements from the personal examination of an orbital chloroma."

What I have found in the specimen under consideration differs from this description in so far only, as the aggregation of colored bodies consisting almost altogether of the globules, (as in the illustration here added), are not mentioned.

What significance, if any, the oxalate of calcium crystals present as to the nature of the disease, I do not venture to say.